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Find the fracture: A case of an occult proximal humerus fracture in a patient with Duchenne muscular dystrophy

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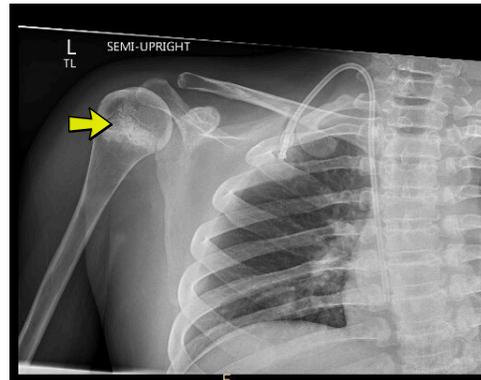
INTRODUCTION

- Duchenne muscular dystrophy (DMD) is an X-linked recessive disorder due to a mutation in the dystrophin gene^{1-2,4-5}.
- Duchenne muscular dystrophy can be characterized by progressive muscular weakness, cardiomyopathy, restricted lung disease, skeletal deformities and cognitive deficits⁵.
- One in 3500-5000 of all males born are affected by DMD¹.
- Chronic steroids are used to help improve muscle strength and neurologic function in patients with DMD, and this increases the risk for osteoporosis¹⁻⁵.
- Patients with Duchenne muscular dystrophy are at an increased risk of fracture due to chronic steroid use, progressive muscle weakness and loss of weight bearing activity¹⁻⁵.

CASE DESCRIPTION

- A 17-year-old boy with a history of Duchenne muscular dystrophy (DMD), non ambulatory and taking deflazacort, presented with a six-day history of right shoulder pain, decreased upper extremity use and ADLs after minor trauma when his wheelchair was not properly tied down on the school bus.
- Bone health history: Last vitamin D level was 58 ng/mL. DEXA scan one year prior had Z scores of -1.6 (height adjust 0.4) for spine, and -3.2, -5.7 and -5.9 (height adjust -2.4, -4.7 and -4.6) for distal femur. He was most recently getting zoledronic acid infusions managed by Endocrinology.
- On initial presentation to the emergency department, his X rays were negative for acute fracture or dislocation in the right shoulder or elbow.
- Exam in rehab clinic:
 - Tenderness along the anterior glenohumeral joint and lateral humerus. There was no tenderness along the clavicle, AC joint or scapular spine.
 - Passive range of motion was restricted to 90 degrees of abduction and 20 degrees of external rotation.
 - His strength was at baseline in the left upper extremity, while the right side had decreased strength from his baseline of 2/5 for shoulder abduction, elbow extension and elbow flexion to 1/5.
 - Special tests including Neer's and Hawkins' were pain limited.
- Further diagnostics: An MRI of the right shoulder revealed a fracture in the metaphysis of the proximal humerus. The fracture was noted to have mild bony compression with extension to the physis and widening of the posterior physis, consistent with a Salter-Harris type 2 fracture.
- Management: Orthopedic surgery recommended wearing a sling for at least three 3 weeks.
- Follow up: After six weeks, he was back to his baseline level of function with no limitations in wheelchair mobility.

X-RAY



AP view of right shoulder showing diffuse osseous demineralization and sclerosis of the proximal humeral physis (yellow arrow).

MRI



Sagittal T1 view of the right shoulder demonstrating a non-displaced transverse metaphyseal fracture (yellow arrow) extending to the physis (Salter-Harris II).

DISCUSSION

- Pediatric proximal humerus fractures are not common⁸.
- Bone health should be addressed at all clinical visits for patients with DMD³.
- Despite a negative x-ray, his broad shoulder and humerus pain, the severity of his functional decline and physical exam prompted further evaluation which revealed an occult fracture on MRI.
- When accessible, MRI is the preferred imaging study to evaluate for occult fracture while also assessing for injury to major soft tissue structures. It has a similar specificity to CT imaging, although it is more sensitive⁹.
- Wheelchair safety is one of the main concerns in this case as the patient did not have adequate wheelchair securement on the bus.
- Per Section 38.23d of the DOT ADA guidelines, wheeled mobility devices should have four straps to tie down the wheelchair in addition to a lap belt and shoulder strap⁶⁻⁷.

CONCLUSIONS

- Physicians caring for patients with DMD on chronic steroid therapy should be aware of increased fracture risk in the upper and lower extremities.
- Fracture should be ruled out if there is high clinical suspicion despite a negative x-ray.
- Physicians, patients, caregivers and transit operators should all be aware of the ADA guidelines for proper wheelchair securement on public transit.

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